

Abnormal cardiac signs after Fontan type of operation: indicators of residua and sequelae

MAURICE P LEUNG,* LEE N BENSON, JEFFREY F SMALLHORN, WILLIAM G WILLIAMS, GEORGE A TRUSLER, ROBERT M FREEDOM

From the Division of Cardiology and Cardiovascular Surgery, Departments of Pediatrics and Surgery, University of Toronto, Faculty of Medicine and Hospital for Sick Children, Toronto, Canada

SUMMARY Among 74 survivors of the Fontan type of operation abnormal cardiac signs were detected in 46 (62%) at postoperative examination. The findings were analysed in relation to the state of the cardiovascular system of these patients. Cyanosis was present in 10 (13.5%) patients. The causes of cyanosis included residual interatrial shunt (six patients), acquired pulmonary arteriovenous fistulas (three patients) and acquired systemic-to-pulmonary vein communication (one patient). Signs of chronic fluid retention were detected in six (8%) patients. In four of them the fluid retention was related to conduit obstruction and in the remaining two it was secondary to severe subaortic stenosis in one and atrioventricular valvar regurgitation in the other. Organic heart murmurs were heard in 29 (39%) patients. The aetiologies of these murmurs were multiple. They included aortic valve regurgitation (eight patients), subaortic stenosis (seven patients), atrioventricular valvar regurgitation (five patients), pulmonary valve regurgitation (five patients), residual Blalock-Taussig shunt (two patients), residual ventricular septal defect (two patients), residual communication in the main pulmonary artery which had been ligated but not divided (one patient), and left ventricular to right atrial shunting (one patient). Cardiac rhythm disturbances of varying aetiology were noted in 23 (31.1%) patients. Sixteen (21%) had supraventricular arrhythmias and seven (9.5%) had conduction abnormalities.

The present review suggests that among survivors of the Fontan type of operation abnormal cardiac signs are indicators of residua or sequelae or both of the native cardiovascular anomalies or surgical procedures.

Atriopulmonary or atrioventricular anastomosis of the Fontan type¹ provides long term palliation in some patients with tricuspid atresia and some with complex forms of cyanotic congenital heart disease.²⁻⁴ The best result is a patient without any cyanosis, no fluid retention, and a quiet heart that can maintain an adequate output at rest and during exercise. After a Fontan type operation, abnormal cardiac signs such as cyanosis, chronic fluid retention, an organic heart murmur or arrhythmia cause concern. We studied these abnormal cardiac signs to see what their relation is to the residua and sequelae of the Fontan type of operation.

Requests for reprints to Professor Robert M Freedom, Department of Paediatric Cardiology, The Hospital for Sick Children, 555 University Avenue, Toronto, Canada.

*Present address: The Grantham Hospital, Hong Kong.

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Patients and methods

Between January 1976 and June 1986 102 patients had a Fontan or a modified Fontan operation at the Hospital for Sick Children in Toronto. Seventy four of the 83 survivors were followed for from two months to 8.6 years (mean 2.6 years). Table 1 lists the cardiac diagnoses in these 74 patients who were aged six months to 23.5 years (mean 10.6 years). Sixty six (89%) had had previous palliative operation to optimise the pulmonary blood flow (table 2). When the Fontan procedure was performed all systemic-to-pulmonary arterial shunts (except the Glenn shunts) were closed surgically. In 34 patients the right atrium to pulmonary artery connection was a direct tissue anastomosis while in six a conduit (three valved and three non-valved) was used. The rudimentary right ventricle was incorporated into the pulmonary cir-

Table 1 Cardiac anatomy of the 74 patients who had undergone a Fontan operation

		Ventriculo-arterial connection		
		Concordant	Discordant	Double outlet
Absent right AV connection (tricuspid atresia)	42	36	6	0
Double inlet ventricle via:	21			
Two AV valves	15	4	9	2
Common AV valve	6	1	2	3
Absent left AV connection	2	1	0	1
Others:	9	4	4	1
TGA + VSD + straddling MV	2			
TGA + VSD + PS + hypo RV	2			
Criss-cross heart + straddling TV	2			
TOF + hypo RV	1			
AV septal defect + PS + hypo RV	1			
DORV + PS + hypo RV	1			

AV, atrioventricular; DORV, double outlet right ventricle; hypo, hypoplastic; MV, mitral valve; PS, pulmonary stenosis; RV, right ventricle; TGA, transposition of great vessels; TOF, tetralogy of Fallot; TV, tricuspid valve; VSD, ventricular septal defect.

cut in 34 other patients, 12 by direct anastomosis and 22 via a valved conduit. Additional procedures included atrioventricular valve annuloplasty (two patients), arterial switch procedure (three patients), enlargement of a restrictive ventricular septal defect (two patients), and creation of an aortopulmonary window (two patients). We reviewed the in hospital and outpatient records as well as the results of cardiac investigations including cross sectional and pulsed Doppler echocardiography, Holter recordings, preoperative and postoperative cardiac catheterisation and cinecardioangiographic data in these 74 patients. All patients had had at least one echocardiographic examination and 38 (51%) patients had 45 catheterisations in the postoperative period. Abnormal cardiac signs after operation were analysed with reference to the findings at operation and the cardiac investigations before and after operation. The clinical courses of these patients were also noted.

Results

Forty six (62%) patients had one or more abnormal cardiac signs: (a) cyanosis, (b) chronic fluid retention (tissue oedema, pleural effusions, and ascites), (c) organic heart murmurs, and (d) arrhythmias. Of the

46 patients who had abnormal cardiac signs only 23 were symptomatic (New York Heart Association class II, 12 patients; class III, eight patients; and class IV, three patients). Five of the patients with symptoms died three months to 7.8 years after the Fontan procedure (three at or soon after replacement of the stenotic conduit, one at insertion of a conduit between the left ventricle and descending aorta for severe subaortic stenosis, and one soon after the Glenn shunt was taken down surgically because of cyanosis secondary to pulmonary arteriovenous fistulas).

CYANOSIS

Cyanosis at rest was detected in 10 (13.5%) patients two days to 2.8 years after operation. In nine of them it was noted within 12 weeks of operation. Their systemic arterial PaO_2 in room air ranged from 23 to 70 mm Hg (mean 44 mm Hg). The causes of cyanosis included residual interatrial shunts (six patients), acquired pulmonary arteriovenous fistulas (three patients) (fig 1a), and communication between the superior intercostal vein and pulmonary vein (one patient) (fig 1b).

Five of the six patients who had residual interatrial shunts underwent six re-operations two days to 7.8 years after the initial Fontan procedure. The remaining patient who had leakage at the suture line of the atrial patch was treated conservatively because he had severe atrioventricular valvar regurgitation and impaired ventricular function. At re-operation two patients were found to have two residual interatrial communications each and the following types of residual interatrial communications were recognised and repaired: a previously missed congenital communication between the coronary sinus and left atrium (four patients) (fig 1c), a second atrial septal defect that was missed at the initial operation (two patients), and a residual defect at the suture line of

Table 2 Palliative procedures done before the Fontan type of operation in the 74 patients studied

Procedure	No
Caval-pulmonary anastomosis:	
Before the Fontan operation	23
At the Fontan operation	9
Waterston's shunt	7
Blalock-Taussig shunt	41
Pott's shunt	9
Pulmonary artery banding	13
Blalock Hanlon septectomy	1
Nil	8

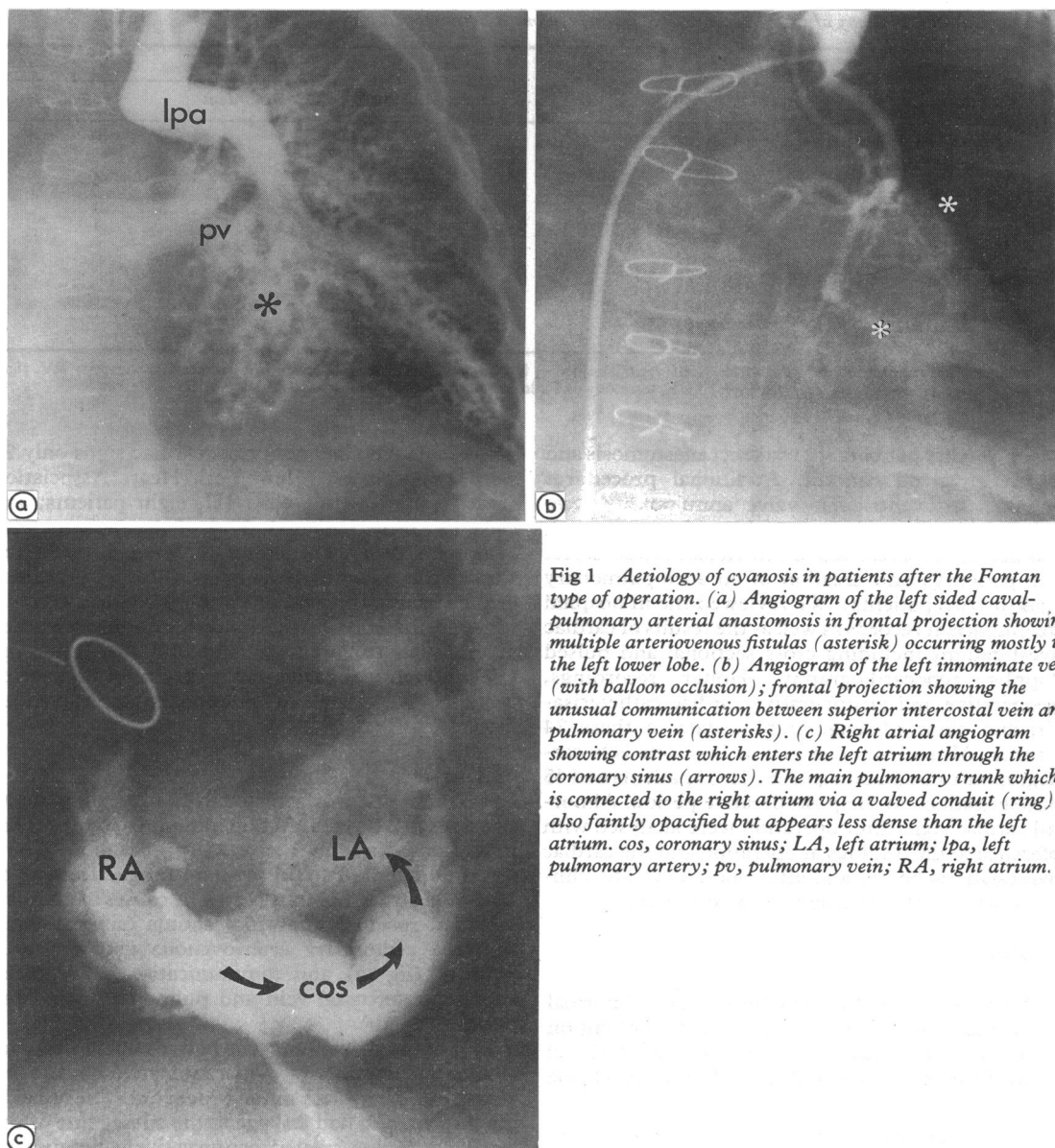


Fig 1 Aetiology of cyanosis in patients after the Fontan type of operation. (a) Angiogram of the left sided caval-pulmonary arterial anastomosis in frontal projection showing multiple arteriovenous fistulas (asterisk) occurring mostly in the left lower lobe. (b) Angiogram of the left innominate vein (with balloon occlusion); frontal projection showing the unusual communication between superior intercostal vein and pulmonary vein (asterisks). (c) Right atrial angiogram showing contrast which enters the left atrium through the coronary sinus (arrows). The main pulmonary trunk which is connected to the right atrium via a valved conduit (ring) is also faintly opacified but appears less dense than the left atrium. cos, coronary sinus; LA, left atrium; lpa, left pulmonary artery; pv, pulmonary vein; RA, right atrium.

the atrial septal patch (one patient). All survived the re-operation except the patient who was re-operated for conduit obstruction 7.8 years after the Fontan procedure and in whom closure of residual interatrial communication was incidental.

In three (9.4%) of the 32 patients who had a Glenn shunt pulmonary arteriovenous fistulas developed six to 17.5 years after the shunting procedure (fig 1a).

Two underwent coil embolisation and one became less cyanotic. The third patient died from pulmonary sepsis after the shunt was taken down surgically.

The cyanosis was mild in the patient in whom an unusual communication developed between the superior intercostal vein and the pulmonary vein (fig 1b). However, he became more cyanotic during an episode of supraventricular tachycardia. He was

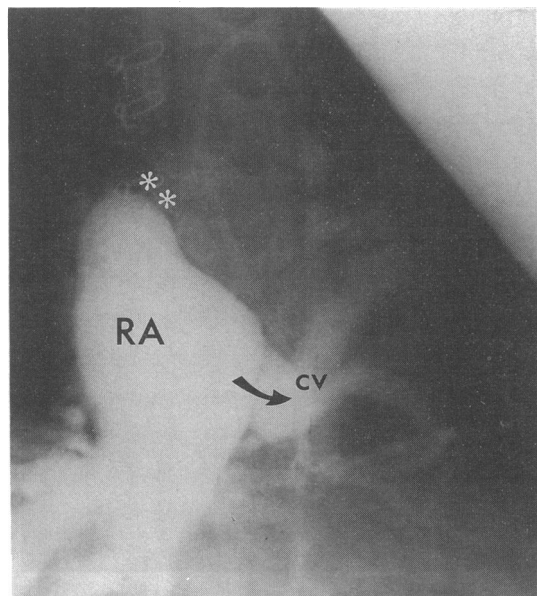


Fig 2 Cause of profound right heart failure in a patient with complete obstruction of the conduit connecting the right atrium to the pulmonary artery. Right atrial angiogram in hepatoclavicular projection showing the absence of forward flow of contrast into the pulmonary trunk (asterisks). There is reflux of contrast into the dilated cardiac veins (arrow) and inferior vena cava. cv, cardiac veins; RA, right atrium.

otherwise well.

CHRONIC FLUID RETENTION

In six (8%) patients persistent oedema, gross hepatomegaly and ascites developed after they had been well for three months to seven years (mean 1.8 years) after the Fontan procedure. In all six patients the mean right atrial pressure was raised (14–27 mm Hg, mean 20 mm Hg) and there was no biochemical evidence of protein losing enteropathy. In four of them there was a coexisting organic heart murmur. The pathogenesis of chronic fluid retention was related to conduit obstruction (fig 2) in four patients (two had an associated mild subaortic stenosis and one had an associated residual interatrial shunt), severe subaortic stenosis (gradient 115 mm Hg) in one other patient, and severe atrioventricular valvar regurgitation in the remaining patient. The last patient was managed conservatively while the other five patients had a repeat operation. Four had conduit replacement, and only one was a long term survivor (one died at re-operation, one each at two months and six months after conduit replacement), and one other patient died at insertion of a valved conduit between the left ventricle and the descending aorta.

ORGANIC HEART MURMURS

Among the 45 patients who did not have an organic heart murmur a soft vibratory functional murmur was heard in 31 (42%) and only 14 (19%) were murmur free. Of the 45 patients 28 (38%) were fully active, without any detectable important residual lesions; eight had arrhythmia; five were cyanotic; one had ascites; three had both arrhythmia and cyanosis; and one had both cyanosis and ascites.

Twenty nine (39%) had one or more organic heart murmurs. We heard 15 long ejection systolic or pansystolic murmurs and 13 early diastolic and three continuous murmurs.

Long ejection systolic (pansystolic) murmur (n = 15)

*With maximum intensity at parasternal areas—*Subaortic stenosis (gradient 10–115 mm Hg) was the underlying cause of the long ejection murmur in seven patients (two had coexisting conduit stenosis, in one other patient there was an associated small shunt from the left ventricle to the right atrium through the surgically “closed” right atrioventricular valve. In three, it was recognised before operation and surgical correction had been attempted. In the remaining four patients it was unmasked after the Fontan procedure (fig 3a). Six of the seven patients had transposition of the great arteries associated with double inlet left ventricle (one with an associated imperforate right atrioventricular valve). The remaining patient had a combination of double inlet and double outlet right ventricle. Only two of these patients had previous pulmonary artery banding. As mentioned above, the three patients with symptoms (two had coexisting conduit obstruction and one had a subaortic gradient of 115 mm Hg) died after re-operation.

A residual ventricular septal defect (Qp:Qs = 1.2:1) was responsible for the pansystolic murmur in two patients with an anastomosis between the right atrium and right ventricle. A residual communication in the main pulmonary artery which had been ligated but not divided was the cause of a high parasternal systolic murmur in one other patient (fig 3b).

*With maximum intensity at the apex—*An apical pansystolic murmur was heard in five patients with a regurgitant atrioventricular valve (common valve, two patients; left sided valve, two patients; right sided valve, one patient). In four patients the regurgitant valve was discovered after the Fontan procedure. Regurgitation was mild and all four patients remained symptom free. In the fifth patient, despite preoperative recognition and surgical repair, the atrioventricular valvar regurgitation remained severe and the patient was in heart failure.

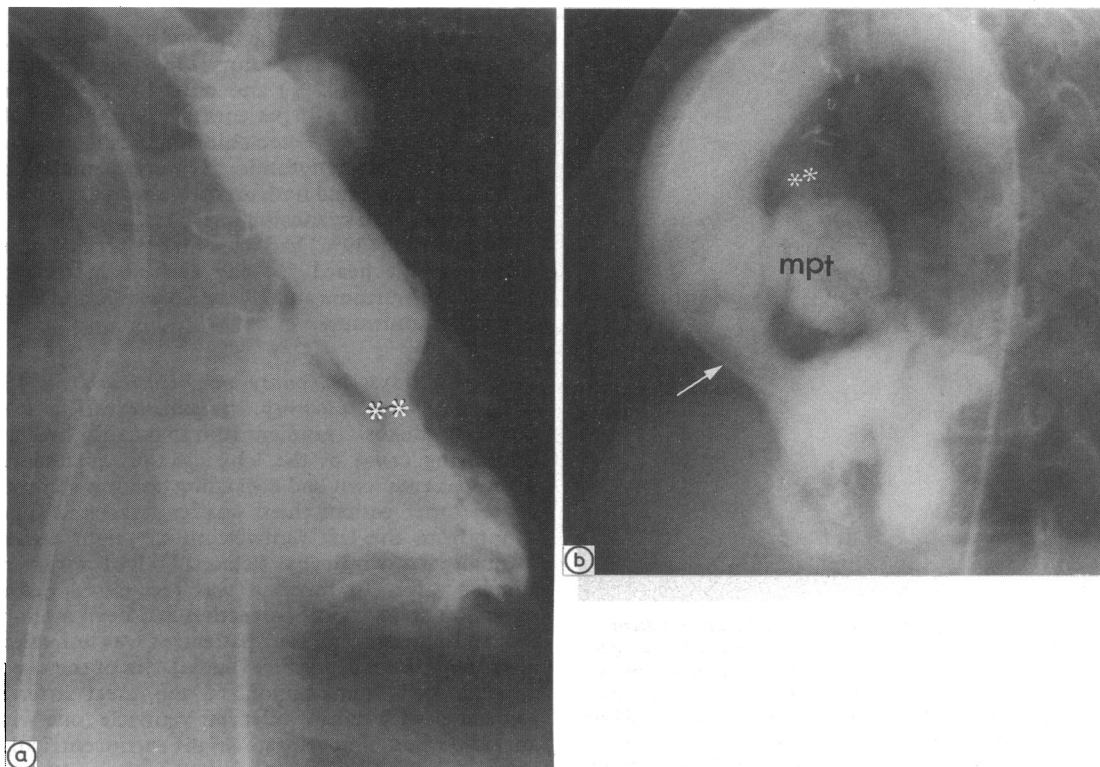


Fig 3 Aetiology of a long systolic or pansystolic murmur in patients after the Fontan type of operation. (a) Left ventriculogram in frontal view showing subaortic stenosis (asterisks) unmasked after Fontan's operation. The patient has transposition of the great arteries associated with a double inlet left ventricle. (b) Left ventriculogram in hepatoclavicular view showing the residual continuity across the surgically ligated (but not divided) main pulmonary trunk (asterisks). The patient has a double outlet right ventricle with mild subaortic narrowing (arrow). mpt, main pulmonary trunk.

Early diastolic murmur (n = 13)

Eight patients had a high pitched early diastolic murmur typical of aortic regurgitation: three had a dilated aortic annulus, three had undergone a concomitant arterial switch procedure (fig 4a), one had a congenital quadricuspid aortic valve, and one had coexistent severe subaortic stenosis. In another five patients with a right atrial-right ventricular anastomosis, a low pitched early diastolic murmur was heard indicating pulmonary regurgitation after insertion of a patch across the annulus of the pulmonary valve.

Continuous murmur (n = 3)

A continuous murmur was heard over the left chest in two patients who had a small residual left Blalock-Taussig shunt (fig 4b). The latter was successfully closed by catheter embolisation in both patients. The third patient with a continuous murmur was awaiting

further investigation.

ARRHYTHMIAS

Atrial rhythm was disturbed in 16 (21%) patients (six patients had associated atrioventricular valvar regurgitation and one other patient had a coexisting communication between the superior intercostal vein and pulmonary vein). Five had more than one form of atrial arrhythmia and in two patients the rhythm disturbance was present before the Fontan procedure. The atrial arrhythmias were supraventricular tachycardia (six patients), atrial flutter (six patients), junctional rhythm (six patients), and wandering pacemaker (three patients). There were no symptoms attributable to atrial arrhythmias except during an episode of supraventricular tachycardia or atrial flutter when patients experienced palpitation and decreased exercise capacity. The cardiac failure became worse in two patients who were in chronic

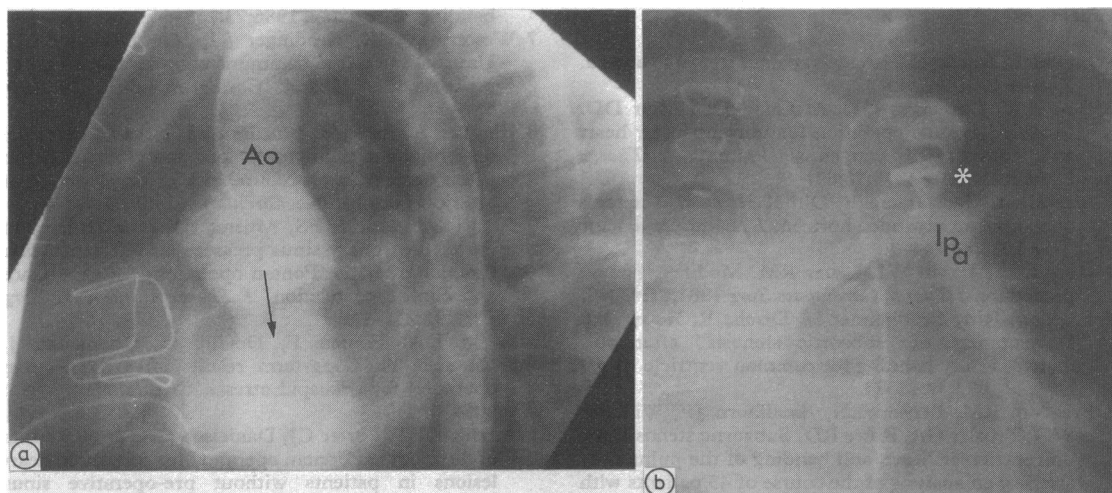


Fig 4 Aetiology of early diastolic and continuous murmurs in patients after the Fontan type of operation. (a) Aortogram in the lateral projection showing moderate aortic regurgitation (arrow) into the main left ventricular chamber. The patient has tricuspid atresia and transposition of great arteries and had undergone an arterial switch operation and modified Fontan's procedure. (b) A left sided Blalock-Taussig shunt persisted after incomplete occlusion by metal clip (asterisk). Ao, aorta; lpa, left pulmonary artery.

heart failure and the cyanosis was increased in the patient who had a coexisting communication between the superior intercostal vein and the pulmonary vein.

Seven (9.5%) patients had complete heart block. In three patients it was present before operation. In the other four patients it was surgically induced during closure of a right sided atrioventricular valve (two patients) or a closure of a large defect in the muscular ventricular septum (one patient) or enlargement of a restrictive ventricular septal defect (one patient). All except one of the patients who were in complete heart block before operation required permanent pacing.

Discussion

We found that most survivors (62%) of the Fontan type of operation had abnormal cardiac signs indicative of residua and sequelae. The commonest abnormal cardiac sign detected was an organic murmur. The pathogenesis of the organic murmur can be deduced readily if the original cardiac anomalies and surgical procedures performed are known. For example, a parasternal pansystolic murmur in a patient who had undergone atrioventricular anastomosis is usually diagnostic of a residual ventricular septal defect. Dysfunctional semilunar and atrioventricular valves and residual systemic to pulmonary artery shunts also have pathognomonic features.

We are surprised that despite a vigilant search

before operation for subaortic stenosis in hearts with a univentricular atrioventricular connection,^{5,6} four patients with this anomaly were not recognised until after the Fontan procedure. In the present study seven (24%) children had organic heart murmurs after operation that were related to subaortic stenosis. When the predisposing anatomy is present, a long systolic murmur after operation may be the first clue to the diagnosis.

The absence of an organic heart murmur, however, does not exclude important residua or sequelae. In our patients, residual interatrial communications and pulmonary arteriovenous fistulas were manifested as cyanosis and important conduit obstruction as chronic fluid retention. The presence of cyanosis after operation precludes long term success and chronic fluid retention is usually also ominous.

Some of the residua and sequelae, such as residual interatrial communications, residual systemic-to-pulmonary artery shunt, and obstruction of the prosthetic conduit are avoidable.⁷⁻¹⁰ But some arrhythmias¹¹ and valve dysfunction³ seem to be inevitable. Further studies to identify these patients at risk and methods to manage these potential lesions are essential if an ideal result is to be achieved in more patients.

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References

- 1 Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971;26:240-8.
- 2 Gale AW, Danielson GK, McGoon DC, Mair DD. Modified Fontan operation for univentricular heart and complicated congenital lesions. *J Thorac Cardiovasc Surg* 1979;78:831-8.
- 3 Marcelletti C, Mazzera E, Olthof H, *et al.* Fontan's operation: an expanded horizon. *J Thorac Cardiovasc Surg* 1980;80:764-9.
- 4 Doty DB, Marvin WJ, Lauer RM. Modified Fontan procedure. *J Thorac Cardiovasc Surg* 1981;81:470-7.
- 5 Freedom RM, Sondheimer H, Dische R, Rowe RD. Development of "subaortic stenosis" after pulmonary artery banding for common ventricle. *Am J Cardiol* 1977;39:78-83.
- 6 Freedom RM, Benson LN, Smallhorn JF, Williams WG, Trusler GA, Rowe RD. Subaortic stenosis, the univentricular heart, and banding of the pulmonary arteries: an analysis of the course of 43 patients with univentricular heart palliated by pulmonary artery banding. *Circulation* 1986;73:758-64.
- 7 Westerman GR, Readinger RI, Van Devanter SH. Unusual interatrial communication after the Fontan procedure. *J Thorac Cardiovasc Surg* 1985;90:627-30.
- 8 Cloutier A, Ash JM, Smallhorn JF, *et al.* Abnormal distribution of pulmonary blood flow after the Glenn shunt or Fontan procedure: risk of development of arteriovenous fistulae. *Circulation* 1985;72:471-9.
- 9 Ilbawi MN, Idriss FS, Muster AJ, *et al.* Effects of elevated coronary sinus pressure on left ventricular function after the Fontan operation. An experience and clinical correlation. *J Thorac Cardiovasc Surg* 1986;92:231-7.
- 10 Girod DA, Fontan F, Deville C, Ottenkamp J, Choussat A. Long-term results after the Fontan operation for tricuspid atresia. *Circulation* 1987;75:605-10.
- 11 Alboliras ET, Porter CJ, Danielson GK, *et al.* Results of the modified Fontan operation for congenital heart lesions in patients without pre-operative sinus rhythm. *J Am Coll Cardiol* 1985;6:228-33.

Leung, Benson, Smallhorn, Williams, Trusler, Freedom